ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 01 | January - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CURRENT KNOWLEDGE IN AETIOPATHOGENESIS AND MANAGEMENT OF BUDD- CHIARI SYNDROME IN PREGNANCY: A CASE SERIES ANALYSIS IN A TERTIARY CARE CENTRE

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ABSTRACT

Budd Chiari Syndrome(BCS) is a congestive hepatopathy due to narrowing and obstruction of hepatic venous outflow. While it is a rare condition, it presents a significant clinical challenge, and pregnancy, because of the prothrombotic state, has a markedly adverse impact on the disease's progression. With the therapeutic advances in medicine and the emergence of new interventional Radiology techniques especially Trans-jugular Intrahepatic Portosystemic Shunt(TIPS), the number of affected women with BCS embarking on pregnancy is on the rise. However, limited evidence exists for the management of pregnancy in such women. Here, we present case reports of 3 patients with BCS to discuss the effect of pregnancy on the course of BCS and vice-versa, and to highlight the necessity of a multidisciplinary approach in such patients. Additionally, we have tried to formulate a management plan for pregnant patients with BCS.

KEYWORDS

Budd Chiari Syndrome, Congestive Hepatopathy, Pregnancy, Prothrombotic State, Multidisciplinary Approach

INTRODUCTION

Budd–Chiari syndrome (BCS) is a rare congestive hepatopathy caused by obstruction of hepatic venous outflow anywhere between the small hepatic veins to the supra-hepatic inferior vena cava (IVC).[1]

BCS has an incidence of 1 in 100,000 in the general population[1]. An underlying hypercoagulable state is responsible for more than 75% of cases, and more than one etiologic factor plays a role in the rest 25%[1].

BCS can be classified as primary or secondary ~~BCS~~ depending on the etiopathogenesis. Primary BCS occurs due to endoluminal venous obstruction due to thrombus and secondary BCS results from compression of hepatic venous outflow tract from extrinsic compression or tumor invasion.

When there is hepatic venous outflow tract obstruction due to blockage of major hepatic veins, the sinusoidal pressure increases, and blood flow decreases. As a result, there is stagnation of interstitial fluid in the liver which causes liver congestion and right upper quadrant pain. ~~And~~ When the capacity of lymphatic drainage is exceeded, the filtrated interstitial fluid escapes through the liver capsule and ascites occur. The portal venous pressure increases and liver perfusion via the portal vein is decreased. As a result, hypoxic damage of hepatocytes occurs with the release of free oxygen radicals. Severe centrilobular hepatic congestion, ischemia, hepatic necrosis, and atrophy occur especially in the perivenular zones. In a few weeks after obstruction, hepatocellular fibrosis occurs, and in a few months, nodular regenerative hyperplasia and cirrhosis develop.

Thus, the clinical presentation is varied and can present as acute/ subacute/ fulminant liver disease or liver failure, or as chronic portal hypertension. Patients may present with acute symptoms like sudden onset abdominal pain, ascites, and hepatomegaly; or have a chronic presentation with symptoms of long-standing portal hypertension.

Interventional portosystemic shunts or the development of portal venous collaterals may improve liver function and delay the cirrhotic process.

BCS usually tends to occur in the third to fifth decades of life and could be due to thrombotic or non-thrombotic obstruction of the hepatic venous outflow. However, typically there would be an underlying prothrombotic disorder like myeloproliferative syndromes, hereditary coagulation factor deficiencies, or antiphospholipid syndromes.

As pregnancy is a prothrombotic state, BCS can flare up in pregnancy despite anticoagulation therapy and such patients may end up with fatal complications like bleeding esophageal varices, fulminant hepatic failure, hepatic encephalopathy, and intractable ascites. Rarely, such patients may even need orthotopic liver transplantation. Though BCS is a rare disease, with the therapeutic advances in medicine and the emergence of new Interventional Radiology techniques, the number of women with BCS embarking on pregnancy is on the rise. Moreover, pregnancy being a prothrombotic state has a detrimental effect on the disease course and limited evidence exists for the management of pregnancy in such women.

Here, we present case reports of 3 patients with BCS to discuss the effect of pregnancy on the course of BCS and vice-versa, and to highlight the necessity of a multidisciplinary approach in such patients.

Case 1

38-year-old 4th gravida with previous 3 missed miscarriages, Antiphospholipid Antibody (APLA) positive, Antinuclear Antibody (ANA) negative and chronic hypertensive on medication came for her antenatal checkup with us.

Diagnosed as a case of Budd Chiari syndrome with Inferior vena cava (IVC) stenosis 5 years back while being evaluated for acute onset severe abdominal pain, abdominal discomfort, and vomiting. Given her very high risk of thromboembolism, IVC stenting was done then. She had a stable course of the disease for almost 3 years.

She also had Immune thrombocytopenic purpura, autoimmune thyroid disease, and Type 2 Diabetes Mellitus. She survived a catastrophic event of pulmonary embolism 2 years prior, when she had an embolus 1.6 x 1.4cm involving her left main pulmonary artery and its bifurcation, following which she developed severe Pulmonary Artery Hypertension (PAH) with mild tricuspid regurgitation, probably chronic thromboembolic PAH, NYHA class 3 and was on therapeutic dose anticoagulants.

Afterwards, she missed her follow-up appointments but returned while

pregnant. Given her high risk thromboembolic disease and severe PAH, she was advised Medical termination of pregnancy (MTP), but she wanted to continue her pregnancy at any cost. She was on long-term warfarin, which was switched to therapeutic low molecular weight heparin(LMWH) once pregnancy was diagnosed.

At 15 weeks, her follow-up scan showed severe fetal growth restriction(FGR), severe oligohydramnios with single deep pocket (SDP) 1cm, global cardiomegaly, dilated right ventricle with muscular ventricular septal defect (VSD), and placentomegaly, with Middle Cerebral Artery Peak Systolic Velocity (MCA PSV) s/o fetal anemia. At this time, she was again recounseled and explained the risks of fetal chromosomal abnormalities especially triploidy , genetic abnormalities, and infections. Because of the guarded fetal prognosis irrespective of etiology and very high maternal risk, a medical board was conducted and she was advised MTP with postabortal karyolyte bobs and DNA storage.

A multidisciplinary team including the primary obstetrician, maternal and fetal medicine specialist, cardiologist, hematologist, rheumatologist, hepatobiliary specialist, endocrinologist, critical care specialist was involved in her care. She underwent suction evacuation with cardiac monitoring and intensivist care. For a short period during the procedure, LMWH was withheld and later restarted. Psychological support and contraceptive advice were also given. She was discharged on long-term anticoagulants, steroids for platelet improvement, antihypertensives, and oral hypoglycemics.

Case 2

31-year-old third gravida with 2 prior miscarriages, diagnosed with chronic liver disease, hepatic vein thrombosis, and Budd Chiari syndrome 12 years back when she was evaluated for progressive abdominal distension. She had complete occlusion of the middle hepatic vein and partial occlusion of the portal vein with portal hypertension. Additionally, she was diagnosed with grade 2 esophageal varices, and further investigation revealed that her Budd-Chiari syndrome was secondary to a factor V Leiden mutation. She was on long term anticoagulant therapy with Warfarin. To rule out associated autoimmune diseases, ANA- IF and APLA were tested which were negative.

After explaining the risks and complications in pregnancy, she decided to continue the pregnancy. During early pregnancy, she was switched over from Warfarin to Injection low molecular weight heparin (LMWH) and low-dose aspirin 150mg.

Her First Trimester Screening was low risk, the targeted anomaly scan showed no anomalies, and serial growth scans were normal. She had iron deficiency anemia, which was corrected with oral iron, and also mild thrombocytopenia.

At 33 weeks, she went into preterm labor and was given tocolysis and antenatal steroids. Heparin injections were stopped at the onset of labor pains. However, she had to be taken for an Emergency Cesarean Section for fetal distress. A live preterm 1.79 kg baby was born to her.

Postoperatively, she was given bridging anticoagulation- LMWH was continued and Warfarin was started simultaneously with INR monitoring. Once the therapeutic INR value was reached, LMWH was discontinued and Warfarin was continued.

However postoperatively on day 7, she developed COVID-19. As literature review showed that heparin injections are better for thromboprophylaxis in acute COVID-19 illness, she was given therapeutic dose LMWH and once better, switched back to Warfarin.

A multidisciplinary team including the primary obstetrician, maternal and fetal medicine specialist, cardiologist, hematologist, rheumatologist, hepatobiliary specialist ensured that she had a safe pregnancy and puerperium.

Case 3

36-year-old second gravida with previous normal delivery, a known case of Budd Chiari syndrome, chronic liver disease with portal hypertension for 5 years (diagnosed when she was evaluated for progressive abdominal distension and pedal edema) on Warfarin, autoimmune hypothyroidism on treatment for 5 years and immune-mediated progressive ascending myeloneuropathy. For progressive and intractable ascites, she underwent main hepatic vein angioplasty and stenting 1 year back and was on long-term anticoagulants. Her ANA and APLA were negative.

After explaining the risks in pregnancy, she also decided to continue the pregnancy. She was switched from Warfarin to LMWH and low dose aspirin 150 mg was started from early pregnancy.

Her First Trimester Screening was low risk, the targeted anomaly scan showed no anomalies. She developed gestational Diabetes Mellitus which was controlled with insulin. Her blood pressure remained normal throughout pregnancy. All her growth scans were normal and she underwent full-term vaginal delivery and delivered a baby of 2.8kg. She was also sterilized postpartum and bridging anticoagulation done postpartum and was discharged on Warfarin.

6 months later, she developed an acute short febrile illness with severe anemia, hepatosplenomegaly, hepato-encephalopathy and pre-renal acute kidney injury and succumbed while being investigated for the cause.

DISCUSSION

Budd Chiari Syndrome (BCS) is defined as obstruction of hepatic venous outflow anywhere between the smaller interlobular veins to terminal portion of IVC. It is a rare congestive hepatopathy with hepatic outflow tract obstruction and has an incidence of 1 in 100,000 to 500,000[1] . Most of the cases occur in the third or the fourth decade with preponderance in females[1].

The first case of Budd Chiari syndrome was reported by Lambron in 1842. George Budd who was an internist at Kings College described the classic triad of abdominal pain, hepatomegaly, and ascites in 1845[2]. It was Hans Chiari in 1895 who described the histopathological features of obliterating endophlebitis of the hepatic veins[2].

The clinical presentation of BCS is varied and can present as acute/ subacute/ fulminant liver disease, liver failure, or as chronic portal hypertension. Patients may present with acute symptoms like sudden onset abdominal pain, ascites, and hepatomegaly; or have a chronic presentation with symptoms of long-standing portal hypertension.

The underlying cause, the location and extent of the thrombus, and functional capacity of the liver determine the clinical course of BCS.

Aetiopathogenesis-

a) Primary Budd Chiari Syndrome- due to endoluminal thrombosis

Hepatic venous outflow tract obstruction occurs most commonly due to thrombosis. Underlying inherited or acquired hypercoagulability state could be identified in 75% of patients, and more than 25% of patients had more than one etiologic factor.[3]

The leading cause of hepatic vein thrombosis is primary myeloproliferative diseases, which is seen in 20% of hepatic venous thrombosis, the prevalence reaching up to 45% when occult myeloproliferative disorders are also included[2]. They include polycythemia vera (10%-40% of cases), essential thrombocythemia and myelofibrosis[2]. Overproduction of erythrocytes, granulocytes & platelets from bone marrow causes sludging and occlusion of hepatic veins.

Hereditary thrombophilias like Factor V Leiden mutation, and Prothrombin gene mutation are found in about 25% and 5% of BCS patients, respectively[3] . BCS is associated with heterozygous factor V Leiden and the G20210A mutation of the prothrombin gene and methylene tetrahydrofolate reductase (MTHFR) gene mutation[3]. BCS is also associated with protein C, protein S and antithrombin 3 deficiency; but as these circulating proteins are synthesized by the liver and their production is reduced in liver dysfunction, it becomes difficult to prove them as a causative factor.

Acquired thrombophilias like Antiphospholipid antibody syndrome are found in about 25% of cases[3] . Hyperhomocysteinemia is also a risk factor for BCS.[3]

Use of oral contraceptives with high estrogen content is also a risk factor for BCS especially when the patients have an underlying inherited or acquired thrombophilia[4]. Similarly, paroxysmal nocturnal

hemoglobinuria(PNH), Behcet's disease, Sjogren's disease, ulcerative colitis, or celiac disease may be a cause of BCS when combined with underlying thrombophilias.[4]

b) Secondary Budd Chiari Syndrome- due to external compression of hepatic veins

Compression caused by neoplasms of adjacent organs like hepatocellular carcinoma, adrenal or renal cell carcinoma, parasitic liver diseases like hydatidiform cysts or amoebic liver abscess, large hepatic nodules are rare etiologic factors causing secondary BCS[5].

Pathological changes include enlarged nutmeg liver with gross venous congestion, sinusoidal congestion, ischemic injury to the liver, and centrilobular necrosis with hemorrhage in the acute stage. While in the chronic stage of BCS, there is fibrosis, scarring, and regenerative nodules with portal hypertension and an enlarged caudate lobe.

Diagnosis

BCS should be suspected in patients with a) sudden onset ascites and painful hepatomegaly; b) massive ascites, but relatively preserved liver functions; c) fulminant hepatic failure along with ascites and hepatomegaly; d) unexplained chronic liver disease; e) liver disease with thrombotic disorder.

Serum transaminase levels may be elevated up to five times the upper limit of normal, especially in the acute and fulminant forms of BCS. Serum alkaline phosphatase and bilirubin levels also increase.

Doppler ultrasound of the liver is the imaging modality of choice when BCS is suspected. It has a sensitivity and specificity of > 85%[6]. Hepatic veins without a flow signal, evidence of hypoechogenic hepatic vein thrombus, multiple collaterals in the hepatic venous circulation, stasis, reversal, or turbulent flow in major hepatic veins or inferior vena cava are indicative of BCS.

Magnetic resonance imaging (MRI) aids in distinguishing between acute and chronic forms of Budd-Chiari syndrome (BCS) by examining the liver for signs of chronic disease, assessing blood flow patterns (which can indicate acute BCS with vessel obstruction), identifying ascites (more common in chronic BCS due to prolonged hepatic venous congestion), and visualizing thrombosis in the hepatic veins and inferior vena cava (fresh thrombi in acute cases and organized or recanalized thrombi in chronic cases). [6]

A Computed tomography (CT) scan may be recommended for better outlining the liver morphology, vascular anatomy, and collateral circulation in the liver especially when a transjugular intrahepatic portosystemic shunt is considered.[6]

Hepatic venography or percutaneous hepatic vein catheterization is the gold standard procedure for the evaluation of hepatic veins and the extent of thrombosis. Spider web pattern in hepatic venography demonstrating rich collaterals is diagnostic of BCS. However, the disadvantage is cannulation of the hepatic vein is difficult and a considerable amount of iodine-containing contrast agent is required. Inferior cavography demonstrates stenosis or occlusion of the inferior vena cava and should be considered if percutaneous or surgical shunts are planned.

A liver biopsy reveals congestion, liver cell necrosis, and fibrosis predominantly in the centrilobular area. The congestion can be seen in heart failure and constrictive pericarditis. Liver biopsy helps to distinguish BCS from veno-occlusive disease, which occurs due to nonthrombotic obstruction of hepatic venules due to sinusoidal wall injury.

Once the diagnosis of BCS is established, a thorough search for the etiology is needed. Complete blood count, peripheral smear, determining of coagulation factors and inhibitors, hereditary and acquired thrombophilia testing, and flow cytometry to detect paroxysmal nocturnal hemoglobinuria should be done. Bone marrow biopsy is necessary if there is a suspicion of primary myelo proliferative disorder.

Management

The management of BCS depends on the underlying cause, the location and extent of the thrombus and the functional capacity of the liver.

Management of BCS involves multidisciplinary care, treating the underlying condition- eg. Eculizumab for treatment of PNH, venesection/hydroxyurea in myeloproliferative disorders, low dose aspirin in essential thrombocythemia. Lifelong anticoagulation is needed which will prevent propagation or recurrence of thrombus but does not reopen the valves.

Endovascular therapies including minimally invasive procedures like percutaneous angioplasty +/- stenting of hepatic veins and inferior vena cava help dilate the stenosis and have proven > 80% patency at 5 years.[7] Transjugular intrahepatic portosystemic shunting (TIPS) establishes an artificial connection within the liver between the inflow portal vein and the outflow hepatic vein. TIPS has lower morbidity and lower mortality rates than open surgery and thus is the preferred approach now. It is the first step in patients who initially present with acute hepatic failure. The primary patency rate using PTFE (polytetrafluoroethylene)-covered stents is 67% at 2-year follow-up, and the 5-year survival rate is >70%.[7]

Liver transplantation is the last resort and is done if the patient worsens despite aggressive medical and radiological treatment. It is the only treatment in fulminant form. TIPS is performed prior to provide additional time during the waiting period for transplantation.. Lifelong anticoagulation is requires following it. A survival rate exceeding 70% at 5 years has been observed, albeit based on limited evidence from a small case series. [8]

Thus treatment options include a step-wise approach with addressing the underlying prothrombotic risk factor, beginning anticoagulation immediately and titrating with anti Xa levels, lifelong anticoagulation, assessing and treating complications of portal hypertension, considering minimally invasive procedures such as thrombolysis, percutaneous angioplasties and transjugular intrahepatic portosystemic shunting (TIPS), and rarely surgical portosystemic shunting in those patients who don't improve. If all these treatments fails/ there is fulminant hepatic failure and end stage liver disease, orthotopic liver transplantation remains the last resort. As very limited data exists, treatment is based on expert consensus only.[6]

BCS in pregnancy

Pregnancy with its prothrombotic state itself is a risk factor for BCS & BCS may be diagnosed for the first time in pregnancy. They may present as an acute disorder with acute major hepatic vein obstruction and rapid deterioration , variceal bleed and even fulminant hepatic failure. Delay in diagnosis or treatment results in irreversible liver damage and high mortality rate.

In patients already diagnosed with BCS, pregnancy can have deleterious effects. Apart from pregnancy-induced hypercoagulability, several inherent changes during pregnancy can negatively impact BCS. These include the prothrombotic state, expansion of plasma and blood volume, hypoalbuminemia, venous stasis, increased intra-abdominal pressure, pressure from the gravid uterus on the inferior vena cava and lymphatic system, and displacement of intra-abdominal organs by the enlarged uterus. The occurrence of thromboembolic events despite full-dose and a Xa-adjusted anticoagulant treatment highlights the high thrombotic risk of patients during pregnancy.[7]

All our patients were diagnosed with BCS prior to pregnancy itself and had atleast one autoimmune disease co-existing, suggesting that there invariably could be an underlying autoimmune basis for developing Budd Chiari Syndrome too.

Preconceptional care

Preconceptional care in BCS patients include multidisciplinary input, counselling of risk associated and ensuring a stable course of the disease before embarking on pregnancy. One-fifth of women with BCS have primary infertility & conceive with treatment.[9] In stable patients with a well-controlled disease, BCS is not a contraindication to pregnancy. However, severe exacerbations can occur in pregnancy. It is suggested that women with known varices or portal hypertension undergo a pre-pregnancy gastroscopy and variceal banding to avoid variceal bleed and its complications.[9]

Fetomaternal effects

Maternal effects include increased risk of intrahepatic cholestasis, pre-eclampsia, abruption, Postpartum hemorrhage.[9] Maternal mortality is not increased in controlled disease, but morbidity is increased.

Due to underlying prothrombotic disorders and placental

malperfusion, fetal outcome maybe poor. Pregnancy loss before 20 weeks is as high as 38%. [7] The risk of preterm labor, prematurity, fetal growth restriction, fetal distress and still birth is also increased.

The clinical course of pregnancy in BCS patients is widely varied. As in our case reports, we had to do Medical Termination of Pregnancy (MTP) for 1 patient due to severe Pulmonary Artery Hypertension (PAH) and severe Fetal Growth Restriction (FGR), 1 patient had preterm labor and delivery at 33 weeks for fetal distress; while the other patient progressed to full term and delivered vaginally with no complications. Likewise, fetal-neonatal outcome was poor, with a high rate of pregnancy losses (2 of our cases had recurrent miscarriages & 1 of them went into preterm labor and delivered a preterm baby)

There was no maternal mortality during pregnancy, delivery and puerperium in our patients with BCS, though one of them later succumbed due to a febrile illness with hepatic encephalopathy. She had succumbed in 6 months following pregnancy. We cannot prove that the deterioration of the course of the disease could have been avoided if the patient had refrained from pregnancy. But the longer time intervals with stable disease and the deteriorating trend during pregnancy and after delivery, however, point to a relevant contribution of pregnancy to the lethal course. More data are needed to adequately manage these challenging cases.

The factors that could influence the outcome of pregnant patients with BCS include whether the disease course was stable before pregnancy, presence and type of prothrombotic factors, number of thrombosed veins, complications of portal hypertension, nature of previous interventions, presence of clinical findings such as ascites, hepatosplenomegaly, esophageal varices, liver function, kidney function, coagulation abnormalities, presence of IVC obstruction and development of collaterals.

Antenatal care

Antenatal care should involve multidisciplinary team approach from the beginning involving maternal fetal medicine specialist, hepatobiliary specialist, hematologist, internal medicine specialist, anaethetist and neonatologist.

A comprehensive history and examination should be undertaken, focusing specifically on eliciting symptoms and signs of a BCS flare. Patients with BCS would be already on anticoagulants; and if on Warfarin, it should be changed to LMWH at the earliest, not later than 6 weeks.

Medical gastroenterology followup every 4 weeks, monthly complete blood count, coagulation profile and liver function tests are warranted.[7] During second trimester, when there is an increased intravascular volume, portal pressure increases and there is an increased risk of variceal bleeding and hence oesophago-gastro-duodenoscopy for variceal surveillance is suggested. If indicated, endoscopic variceal ligation is the treatment of choice in pregnancy. In patients who have undergone TIPS, regular abdominal ultrasound is suggested every 4 weeks to ensure patency.

2 weekly antenatal care till 34 weeks and weekly thereafter is suggested. Serial growth scans from 24 weeks of pregnancy helps identify fetal growth restriction.[7]

A birth plan should be made by 32 to 34 weeks with an MDT input.[7]

Intrapartum care

Therapeutic LMWH is stopped 24hrs prior & UFH 12hrs prior to planned induction or regional anaesthesia.

The aim is to achieve a vaginal delivery, with continuous fetal monitoring during labor and 1:1 care. Cesarean section is indicated only for obstetric indications or if there are large esophageal varices with risk of bleeding.

Senior obstetricians, senior anaesthetists should be involved in Caesarean section of BCS patients especially if they have portal hypertension as there can be large pelvic vein collaterals and chance of postoperative ascites. If deemed necessary, vascular surgeons also should be involved. Succinylcholine may be avoided for intubation, as both pregnancy and hepatic failure are associated with pseudocholinesterase deficiency and an unpredictable duration of neuromuscular block.

Postpartum care

Adequate hydration, early mobilisation, DVT stockings should be encouraged. Therapeutic anticoagulation is restarted 6hours after vaginal delivery and 12hours after Cesarean section. LMWH is overlapped with warfarin until INR 2-3 is achieved. Postnatal gastroenterology and hematology followup also should be done before discharge. Contraceptive advice should be given avoiding estrogen containing pills.

To summarise, based on our clinical experience, we suggest the following recommendations for management of BCS in pregnancy-

- Preconceptional counselling should include risks of BCS in pregnancy- chance of flare-ups, worsening portal hypertension, liver function deterioration, hepatic encephalopathy, increased thrombotic risks; all of which could be lifethreatening. Women should be allowed to enter into pregnancy only after knowing their risks.
- Development of pulmonary artery hypertension in BCS patients is a contraindication to pregnancy in view of very high maternal mortality rates.
- Offer endoscopic screening for varices in patients with portal hypertension, when conception is planned and during the second trimester
- Multidisciplinary team approach from the beginning involving maternal fetal medicine specialist, hepatobiliary specialist, hematologist, internal medicine specialist, anaethetist and neonatologist
- Early pregnancy switch over from Warfarin to LMWH should be considered in view of fetal teratogenicity
- As BCS patients are at high risk of thrombosis, therapeutic dose of LMWH should be considered. In those at very high risk of thrombosis, Warfarin can be continued after the period of organogenesis.
- In addition to LMWH/ Warfarin, adequate hydration and mobilisation should be encouraged in pregnancy to avoid thrombotic risks.
- Close monitoring of hepatic function and high index of suspicion for development of complications
- Serial growth monitoring every 3 weeks from 24weeks
- Vaginal delivery is the preferred mode of delivery (with continuous fetal monitoring during labor and 1:1 care) unless there are severe esophageal varices.
- Stop LMWH therapeutic dose 24hrs prior to delivery. Consider switching to unfractionated heparin(UH) at 36wks (as UH has short t1/2, rapidly reversible action) especially in patients at high risk of thrombosis. If using UH therapeutic dose, stop 12hrs prior to delivery.
- Succinylcholine may be avoided for intubation, as both pregnancy and hepatic failure are associated with pseudocholinesterase deficiency and an unpredictable duration of neuromuscular block.
- Postnatally, adequate hydration and mobilisation is encouraged.
- Can restart LMWH 6 hrs after normal delivery and 12hrs following Cesarean Section
- Postnatally, do bridging anticoagulation from LMWH to Warfarin with daily INR monitoring.
- Avoid estrogen containing hormonal contraceptives as they are at high risk of thrombosis.
- Specialist followup- gastroenterologist/ hematologist prior to discharge

CONCLUSION

To conclude, the management of pregnancy in patients with BCS is a demanding endeavor that poses significant challenges for obstetricians. It necessitates a multidisciplinary team approach within a tertiary care center equipped with ample resources and meticulous monitoring for potential complications. Further studies are imperative to formulate an evidence-based management strategy for patients with Budd-Chiari syndrome.

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